

A PANDAS Story

Published by Steven Novella under Neuroscience/Mental Health

Comments: 61

One of the skills I try to teach medical students on their journey to becoming experience clinicians is to consider and address the patient's "narrative." Patients have a certain understanding of their illness, its cause, and its role in their life. They make sense of their situation as best as they can, resulting in a story they tell themselves. This is how humans generally deal with the complexities of life.

There is a potential problem when the clinical narrative of the health care provider conflicts significantly with the illness narrative of the patient. Patients, for example, often feel that a highly specific diagnosis is necessary for optimal treatment of their condition. Until they are given such a diagnosis they feel they need to keep looking – for better diagnostic tests or different specialists (what I call the "Dr. House" narrative). The appropriate diagnostic and therapeutic algorithm for that patient, however, may not require a specific diagnosis, but rather eliminating certain diagnoses and then treating the probable category or clinical syndrome that remains. The clinical narrative, in other words, may be one of considering risks vs benefits with incomplete and imperfect knowledge.

Increasingly, it seems, the conflict of narratives is taking on a larger scale – not just between one doctor and one patient, but between the medical community and patient communities. Perhaps this is one manifestation of the new social media generation. Take, for example, a recent controversy over a patient diagnosed with PANDAS (pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections).

What is uncontroversial about this story is that Elizabeth Wray is a 16 year old girl who developed the rapid onset of some form of tic or obsessive compulsive disorder. She was diagnosed with PANDAS and treated, for a time, with antibiotics (for the presumed Strep infection). She was recently transferred to Boston Children's Hospital (BCH) for treatment, but her doctors there apparently disagreed with her previous management. They petitioned the state to take custody of Elizabeth partly so that they can have her transferred to a locked psychiatric unit. A judge recent decided that the state of Massachusetts would take temporary custody of Elizabeth while the case is being sorted out, but did not allow her transfer to the locked unit.

As far as I can tell, those are the basic facts of the case. However these facts are woven into very different narratives. Part of the PANDAS patient community has taken up the call to free Elizabeth Wray. Most are quoting the family's attorney, such as this report:

"Their daughter Elizabeth was admitted to BCH due to eating issues. This is the second family I have represented within two months regarding BCH with the same fact pattern. In both cases, BCH has immediately told the parents that PANDAS/PANS does not exist, cut off the child's antibiotics, reported the parents to Mass child protective services, and had the parents trailed by guards at the hospital. In both cases, BCH has actively encouraged children's services to remove the child from the custody of the parents unless the family signed a voluntary agreement to place the child in a locked psych unit."

This is sickening. So very sickening.

Please forward this to everyone you know. Parents who are dealing with PANDAS are going through so much. To have their child taken from them, by **ignorant** doctors who claim the condition doesn't even exist?

According to this narrative, PANDAS is a very real syndrome that affects many children and responds to treatment. However, most doctors are dismissive, ignorant, and arrogant and "just don't get it," so they deny the existence of PANDAS and as a result refuse needed treatment to desperate patients. There are a few mavericks willing to buck the system for their patients, and they are champions of a populist crusade against entrenched professional ignorance. Sometimes insurance companies also get to wear the black hats, or Big Pharma, or government regulators, or all three.

It's a great story, complete with villains and heroes, and has at its core the hope of a treatment for a serious and frightening illness. It is perhaps naive to think that this powerful narrative can be addressed by something as cold and dry as scientific evidence. The narrative, in fact, shields itself from such evidence.

The medical community, as you might imagine, has a very different narrative, one that you will read here on Science-Based Medicine quite often (yes, we have our own narratives that guide our thinking about often complex topics). According to the SBM or mainstream narrative, medicine is a complex business. It is especially difficult to identify new diseases and clinical entities. The creation of a new diagnosis must be done very carefully, with thoughtful scientific studies used in an open-minded way to explore all the complexities involved. Only when all the ducks are in a row can we confidently conclude that a new diagnosis is, in fact, a distinct entity, and not just a variant or misidentification of existing known diseases. Meanwhile, even before all the diagnostic questions can be answered, we need research to answer the basic question of how patients respond to specific treatments – what clinical features predict what kind of respond to which treatments? This is, as I alluded to above, not necessarily the same question as one of diagnosis.

Often new ideas in medicine are controversial for years, even decades, before a consensus can be reached about this fundamental claims. And most new ideas in medicine turn out to be wrong, but slowly our medical knowledge grinds forward.

How does PANDAS relate to the above narrative? A recent review found:

The relationship between obsessive-compulsive disorder (OCD) or tics/Tourette's syndrome in childhood to antecedent group A streptococci (GAS) is unclear. One recent prospective cohort study found that more than 85% of clinical exacerbations in OCD/tic behavior in patients who met criteria for PANDAS had no relationship to GAS infection. Another study found no correlation between clinical exacerbations and changes in a variety of markers of brain autoimmunity, the proposed pathogenesis of PANDAS. A third recent study concluded that, compared with specialty clinic diagnoses, patients diagnosed with tics or Tourette's by physicians in the community were significantly more likely to be diagnosed with PANDAS without meeting the proposed criteria, most lacked supporting laboratory evidence of GAS infection, and they were more likely to be treated with unjustified short-term to chronic antibiotic and/or immunomodulatory therapy.

Despite continued research in the field, the relationship between GAS and specific neuropsychiatric disorders (PANDAS) remains elusive. It is possible that GAS infection may be but one of the many stressors that can exacerbate tic/Tourette's or OCD in a subset of such patients.

I have to stress that I am not an expert in this area, but reading as much of the published literature as I can, the above is essentially what I found. The syndrome of sudden onset neuropsychiatric symptoms certainly exists, and is reported in the literature. However, the pathogenesis is unclear – specifically its relationship to GAS. GAS may not be a specific cause, it may just be one of many possible triggers of an exacerbation of a syndrome that has another underlying cause or causes. Further, the more specific question of whether or not patients who meet diagnostic criteria for PANDAS respond to antibiotics has not been settled, but certainly those advocating for such treatment have not met their burden of proof that it is safe and effective. One researcher, Singer, has proposed moving to a new diagnostic label, CANS (childhood acute neuropsychiatric syndrome), which has the advantage of not making any assumptions about pathophysiology that are as yet unproven.

The bottom line is that PANDAS remains uncertain and controversial. It's not implausible and I (I think like most clinicians) remain open minded about this disorder and are just waiting for better evidence to settle the issue. We're happy to listen to whatever the evidence has to say.

It's difficult to express all of this complexity to a community of parents of children with a scary neuropsychiatric disorder who are just looking for treatment. The internet narrative of heroes and villains makes the job that much more difficult. BCH seems to be caught in the middle of this controversy. They are not allowed to talk about this case, but a statement from BCH has stated that they do treat PANDAS. This suggests, as I suspected, the notion that they categorically deny the existence of PANDAS is a gross oversimplification of their position – suitable for a comic book villain, but not the complexities of modern health care.

Conclusion

Confronting common narrative in the public is critically important for proponents of science-based medicine. I do think this gulf can be bridged, mainly because there is vast common ground. Patient, their families, and the medical community do all want the same thing in the end – to do what is best for patients. Further there is still vast agreement that the scientific evidence is ultimately the best guide to what is best for patients. If we can agree to this common ground, then a much more productive discussion can be had about what the current evidence says, what further studies are needed, and what is the optimum management at present given our state of knowledge.